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"CLOACAL MEMBRANE ANOMALIES" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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ABSTRACT

Cloacal malformation is a rare but important anomaly. The cloacal anomaly is characterised by the persistence of a common channel draining the urinary, genital and alimentary tracts through a single orifice. It results from abnormal compartmentalization of features that are normal in the primitive female embryo. Abnormal embryology and cloacal anatomy are described in detail. Cloacal abnormalities are usually diagnosed promptly in the neonatal period.

Introduction

Abnormal cloacal development takes place when rectum, vagina and lower urinary tract fuse into a single common channel. Persistent cloaca is a most severe malformation of cloacal anomalies in girls and is associated with complex pelvic malformations. The abnormality of these structures varies from bladder neck to just beneath the perineal skin. In rare cases, the structures converge in the bladder with a cleft or with only rudiment attachment. Generally, the length of the common channel correlates well with the sphincter mechanism of the anus and the bladder. Different patterns of the cloacal anomaly result from differences in the level of arrest in the early development of urorectal septum. The position of rectal fistula may cause the vaginal anomalies. The müllerian ducts don't fuse in the existence of high rectal fistula and results in duplication of the uterus and the vagina. Anorectal malformations are not uncommon. A child who is born with one presents soon after birth with abdominal distension and the failure to pass meconium. Some lesions are not complete, and present later with difficulty passing faeces, or distension. There are many kinds of lesion, but what matters most is whether a child's rectum ends above or below his puborectalis sling. If the rectum extends below it, you can make him an anus, or dilate a stenosed one without difficulty. But if his rectum ends above this sling, you will have to make a temporary colostomy, and refer him later for his sigmoid colon to be pulled through to his perineum. This is difficult, but continence can be achieved in most cases.

A child's anus or rectum sometimes fail to develop entirely (agenesis), they may partly fail to develop (atresia), or his rectum or anus may be narrowed (stenosis). Agenesis (but not the other lesions) may be combined with fistulae between the rectum, and the urinary, or genital tracts, of either sex.

These variables combine to produce a complex series of lesions. Some fistulae are useful, because you may be able to dilate them to make an anus.. Initial management focuses on anatomic remodelling of the urinary and gastrointestinal system to achieve continence. Improved paediatric management strategies have increased the patient survival into adult life. In order to provide appropriate advice, clinicians who are undertaking life-long management of adolescent and adult patients need to understand functional outcomes, as well as sexual function and reproductive potential.

Incidence

The reports states that the incidence rate of cloacal malformations is approximately 1 per 20,000-25,000 live births. The records often contain reports of rectovaginal fistula, imperforate anus, anal agenesis and other abnormalities. Anorectal malformations occur in approximately 1 per 5000 live births

Normal fate of cloacal membrane:

The hindgut follows the midgut, in the embryo, and extends from the posterior intestinal portal to the cloacal membrane. It gives rise to the left one-third to one-half or distal portion of the transverse colon, the descending colon, the sigmoid or pelvic colon, the rectum, the upper portion of the anal canal, and part of the urogenital system (e.g., the bladder and urethra). It is supplied by the inferior mesenteric artery. The terminal part of the hindgut enters into the cloaca, which is an endoderm-lined cavity that is in direct contact with the surface ectoderm. The cloacal membrane is composed of endoderm of the cloaca and ectoderm of the proctoderm or anal pit. The cloaca, receives the allantois ventrally and the mesonephric ducts laterally

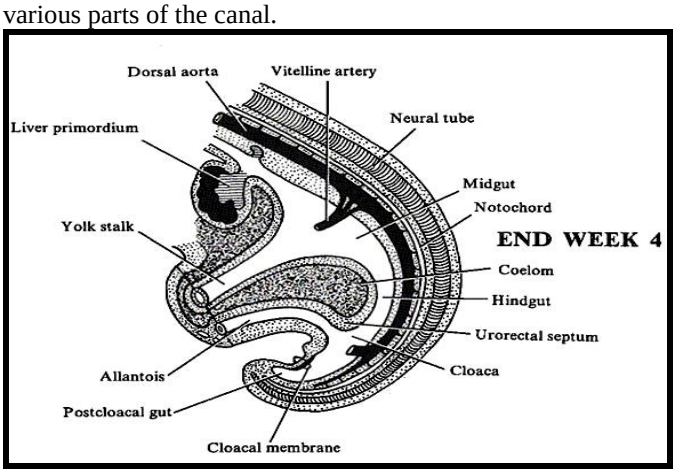
Partitioning of the cloaca

During normal development, a coronal wedge or ridge of mesenchyme, the *urorectal septum*, forms in the angle between the allantois and the hindgut As the septum grows caudal toward the cloacal membrane, it divides the cloaca into an anterior portion, the *primitive urogenital sinus*, and a

posterior part, the *anorectal canal***.** By seventh week, the urorectal septum reaches the cloacal membrane and fuses with it. Thus, the membrane is divided into a posterior *anal membrane* and a larger anterior *urogenital membrane*. The area of fusion of the urorectal septum and the cloacal membrane becomes the *primitive perineum or perineal body*. During eighth week, proliferation of mesenchyme around the anal membrane raises the surrounding ectoderm to form a shallow pit, the *anal pit or proctodeum*. The surrounding swellings are called the *anal folds*. The anal membrane usually ruptures at the end of the eighth week, bringing the distal part of the digestive tract (anal canal)into communication into the amionitc cavity.

The anal canal

Its upper two-thirds is derived from hindgut and its lower third from the proctodeum or anal pit. The junction of the anal pit ectoderm and the hindgut endoderm is indicated by the *anatomic anorectal or pectinate (dentate) line* which is at the level of the anal or semilunar valves. This is the former site of the anal membrane and where the epithelium changes from columnar to stratified squamous. At the Anus, the epithelium is keratinized and continuous with the surface skin of the perineum. The surrounding tissue is derived from splanchnic mesenchyme or mesoderm. The hindgut part of the anal canal is supplied by the inferior mesenteric artery; whereas, the power portion (anal pit) is supplied by the internal pudendal branch of the internal iliac artery. As the result, the difference of the embryological origin of the upper and lower parts of the anal canal, venous (external and internal hemorrhoidal or rectal plexi) and lymphatic drainage and nerve supply (autonomic and peripheral) differs in the various parts of the canal.



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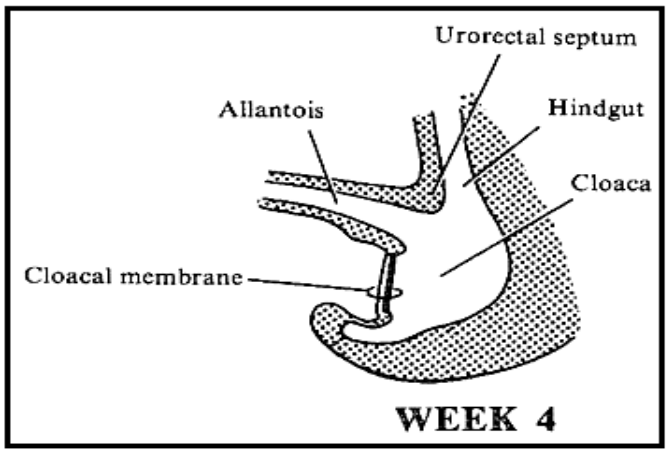

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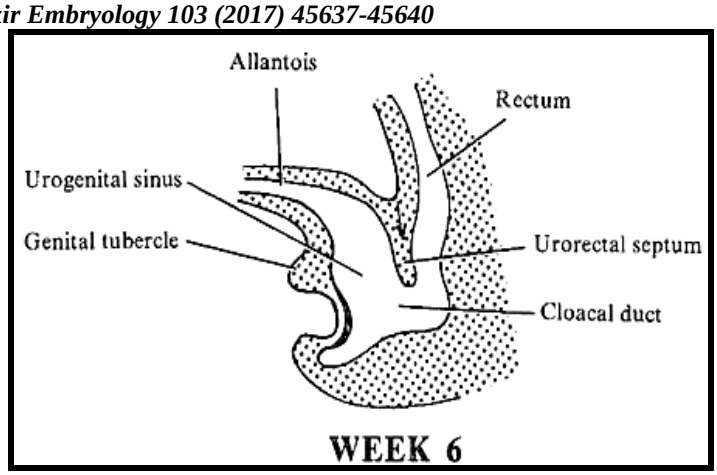

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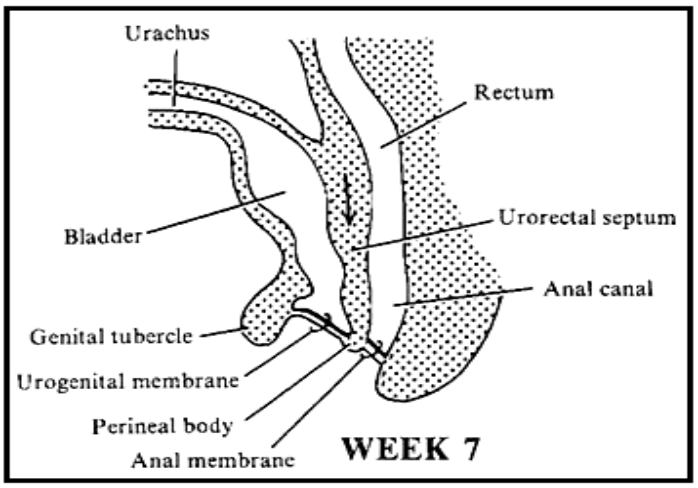

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Fig1. A, B, C, D Schematic representation shows the development of rectum and anus from hindgut.

Ontogenesis of abnormal development of cloacal membrane

Exstrophy of the bladder

Typically, after 4 weeks of life, the urorectal septum divides the cloaca into an anterior urogenital sinus and a posteroanorectal canal. Simultaneously, the cloacal membrane is invaded by lateral mesodermal folds at approximately 4 weeks of gestation. It is postulated that if a mesoderm invasion does not occur, the infraumbilical cloacal membrane persists, along with poor abdominal wall development. Because of its inherent instability, the cloacal membrane eventually ruptures. If it does so before the urorectal septum descends at 6 to 8 weeks of gestation, then cloacal exstrophy results. However, in the developing embryo, a stage similar in appearance to cloacal exstrophy does not exist. Therefore, the anomaly must not represent an arrest in development, but more likely some form of embryo genetic defect. According to Muecke (1964), an abnormally extensive cloacal membrane produces a wedge effect, serving as a mechanical barrier to mesodermal migration, which results in impaired development of the abdominal wall, failure of fusion of the paired genital tubercles, and diastasis of pubis. Exstrophy of the cloaca results when the wedge effect occurs before the formation of an urorectal septum at 6 weeks.

Retrourinary Fistula

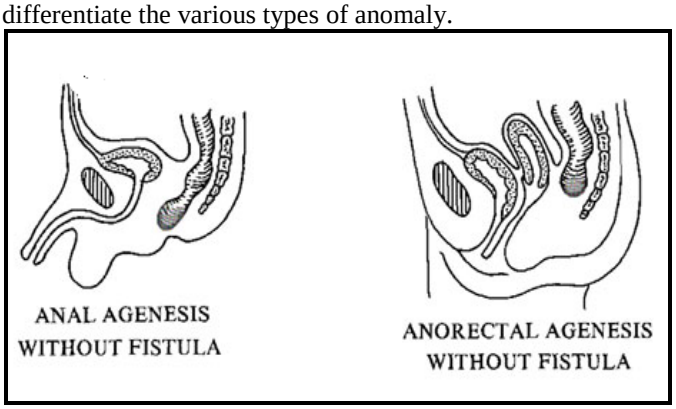
This is one of the anomalies which occur by connecting rectum and lower urinary tract (rectovesical and recto urethral fistulas, etc.) or vagina. Due to the unusual division of cloaca into rectum and the urogenital sinus.

A recto-bladder neck fistula, sometimes referred to as a rectovesical fistula, is one of the least common Anal-Rectal Malformations seen in male patients. The condition is characterized by a connection between the rectum and the bladder, which can cause urine and feces to mix and empty out of the urethra. A recto-urethral fistula is characterized by the abnormal connection of the urethra to either the anus or rectum. This results in both solid waste and urine emptying from the body through the urethra.

Anal Agenesis, with or without Fistula

The anal canal may abruptly end without a orifice, or may be with an ectopic anus or an anoperineal fistula that opens into the perineum. This occurs due to the incomplete separation of the cloaca by the urorectal septum.

Anorectal agenesis with or without fistula: rectum ends blindly above the anal canal, but there is usually a fistula to the urethra in the male or vagina in the female. Pure rectal atresia: complete failure of the formation of the inferior part of the rectum and anal canal Rectal atresia with fistula: always insufficient. The length and degree of anastomosis differentiate the various types of anomaly.



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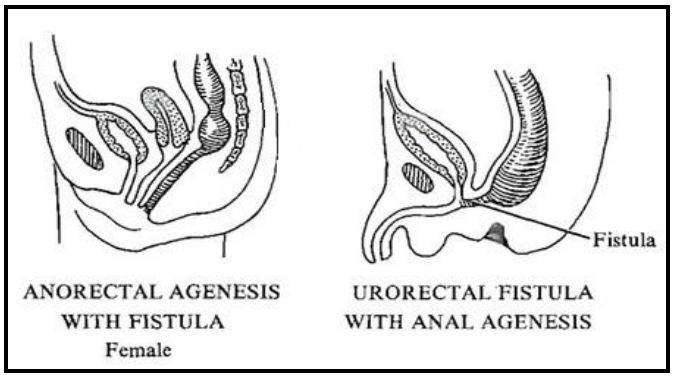

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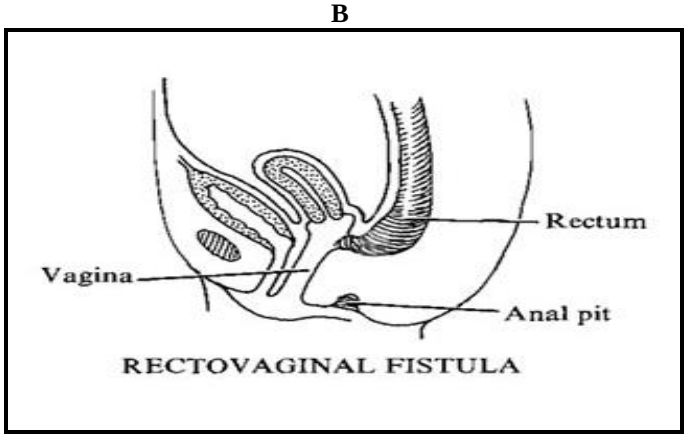

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Fig 2. A, B, C Schematic representation shows the different type of anorectal anomalies.

Discussion

A cloacal anomaly, occurs because of the incomplete urorectal division because of an arrest in the descent of the urorectal septum. There are different types of anomaly according to the levels of arrest, with a high arrest developing a long common cloacal channel. The confluence may be at the bladder neck or even in the bladder. The length of the common channel correlates well with the sphincter mechanism of the anus and the bladder. The frequency and severity of genitourinary abnormalities may be related to the level of the confluence.

A high position of the rectal fistula would prevent fusion of the müllerian ducts followed by duplication of the uterus and the vagina, or even vaginal hypoplasia. At birth, any girl with a single perineal orifice requires abdominal ultrasonography to evaluate the kidneys and the urinary tracts as well as the association of vaginal dilatation. The delayed diagnosis of the anorectal malformation has the potential to contribute to unnecessary patient suffering and in our experience diminished parental confidence in their physicians. It is important to be aware that not all anorectal malformations will present in the newborn period as an imperforate anus. Examination of the anal region in the newborn should involve more than just confirming the presence of an anal orifice. Attention should be taken to note the location and appearance of the anus. Clinical suspicion should be heightened in infants or toddlers who present with an exacerbation of constipation associated with a recent change in diet. Any suspicion of an abnormally located anal opening, regardless of its subtlety, warrants referral to a surgeon with expertise in managing anorectal malformations. Rectal atresia has been treated by pulling the blind proximal bowel through the distal pouch; the treatment of cloaca has varied based on the anatomical abnormality. There are also more number of anomalies like, Imperforate anus is a defect that is present from birth The opening to the anus is missing or blocked. The anus is the opening to the rectum through which stools leave the body. The infant should be checked for other problems, such as abnormalities of the genitals, urinary tract, and spine .Surgery to correct the defect is needed. If the rectum connects with other organs, these organs will also need to be repaired. A temporary colostomy (connecting the end of the large intestine to the abdomen wall so that stool can be collected in a bag) is often needed. Rectovestibular fistula is the most common type of anomaly found in a female newborn with anorectal malformation. Müllerian agenesis or Rectovestibular fistula is a congenital malformation of the genital tract. The embryological origin of the genital tract is derived initially from the intermediate mesoderm of the urogenital ridge. Müllerian agenesis has serious implications on the child's eventual ability to reproduce. Early diagnosis of this condition allows the psychological issues and concerns to be carefully evaluated and addressed earlier in the child's life.

Cloacal anomalies in girls are one of the most complex and challenging reconstructions of the genitourinary systems. The anatomical variety of this entity determines the management options from in utero. It involves not only the creation of three orifices in the perineum, but also stable renal function and a continent catheterizable urethra. Accomplishment of the definitive repair requires the combined expertise of experienced pediatric surgeons and pediatric urologists.

Conclusion

As it is said in this article, cloacal membrane formation is one among the most important formation in a developing foetus. Eventhough there are only some of the anomalies mentioned here, there are a lot of them we haven't discussed yet. At last I would like to thank Mr. Ganesh Elumalai, for givig me this opportunity to see about these various complications of cloacal membrane.

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